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SCLERODERMA.[1]

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IT is not without hesitation that I bring before you this evening the subject of scleroderma; first, because the relations of this disease to the nervous system have not been clearly defined, and secondly, because the name itself implies that the subject properly belongs to the field of dermatology. However, we may find some justification for treating this subject from the neurological standpoint, when we reflect that such diseases as myxœdema and acromegaly have been tacitly admitted as falling within the sphere of the neurologist, and further that various considerations, both clinical and pathological, suggest that scleroderma belongs to the interesting group of trophic affections of which the diseases just mentioned, myxœdema and acromegaly, constitute such conspicuous examples.

Before entering into a discussion of the position and relations of scleroderma, however, I propose to detail before you, briefly, the histories of three cases that have been under my observation. Following this a brief analysis of the subject will be attempted. Let me preface the account of these cases by the general statement that probably in the majority, if not all of the cases of scleroderma, two, and even three stages, can be distinguished. Unquestionably, this is true of diffuse scleroderma, and I believe that it is equally true, though not so readily demonstratable, for the circumscribed and more purely localised forms. In the diffuse form there is early noted a condition in which the substance of the derma, and it may be of other structures, is increased in both volume and density. It is true that cases can be separated into two groups, according to the physical qualities of this enlargement; namely, into the indurated and the œdematous forms. Even a third form is recognized in which the volume of the skin is not so much affected as is its density and its resistance. However, whatever view be adopted, we are justified, tentatively at least, in speaking of an early or first stage of the affection, a stage which in typical diffuse scleroderma is characterized by infiltration and enlargement. I will begin by briefly detailing the history of such a case.

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[1] Read before the Academy of Medicine of New York, April 16, 1896.

Case I, C. S., aged 43 years, a German, a machinist by occupation, while in good health met with the following accident on August 4, 1894. While in a position in which his head was strongly inclined forward, a piece of ice, weighing about twenty pounds, fell from a height of some fourteen feet, striking him upon the back of the head. He received a severe transverse lacerated wound over the occiput, severing all of the structures of the scalp. He was unconscious for a few seconds, and was subsequently taken to the Jefferson Hospital, where the wound was treated.

He remained at the hospital for four or five days. Though the wound healed promptly he continued to suffer severely from the shock of the accident, and also from severe occipital headache. It was for this headache that he was referred to the Nervous Department of the hospital. With the exception of a rather ragged scar the physical examination proved negative. There was no alteration of reflexes, no change in the eye-grounds, nothing but severe pain referred to the site of the injury. Some tenderness was also noted in the scar and adjacent structures, but this subsequently disappeared. I was inclined at this time to regard the case as one in which injury of the bone, possibly a fracture of the internal table had occurred, or more probable still, that the blow upon the head had resulted in a localised traumatic pachymeningitis. He reported at the Out Patient Department at irregular intervals until the fall of 1894 Not having seen him for some time I now noticed that his appearace had decidedly changed. His face seemed much fuller, and when I began as before to examine the scar over the occiput, I noted to my great astonishment, that the neck was swollen. I at once stripped him, and now observed that considerable swelling was noticeable over the shoulders and arms, and to a less extent over the forearms. There was also an unmistakable swelling of the skin over the chest, abdomen and back, and even to some extent over the thighs and buttocks. My first thought was of myxœdema, but not only the facial expression but the resistance offered by the skin differed markedly from that which I had seen in the last mentioned disease. The character of the hair, the shape of the hands, which were normal, also forbade such a diagnosis. The coarser folds of the features were preserved, but many of the smaller wrinkles were smoothed out and effaced. When questioned he stated that the pain in his head had persisted ever since the accident, but that between two and three months afterward he noticed that the skin on the back of the neck, and about the scar in the occipital region, was stiff and hard. This stiffness spread gradually around the neck and face, and then downwards over the shoulders, arms and trunk. The swelling had at the same time, become gradually more and more pronounced. In addition to the occipital pain already mentioned, the man complained of marked general weakness, and was excessively depressed in spirits. His knee jerks were markedly plus. Station and gait revealed no peculiarities. There were no sensory disturbances. Pigmentary changes were not noted in the skin at this time. The induration gradually increased until it involved distinctly all portions of the body, though the legs and feet were never affected to a marked degree.

An examination of the thoracic and abdominal viscera revealed nothing abnormal. A qualitative examination of the urine proved negative. The specific gravity of the urine, however, was high (1028) and when examined quantitatively, it was found to contain an unusual proportion of solids. It appeared to be merely a concentrated urine with the exception of the phosphates, which were much in excess. Efforts were made to get the patient to measure the urine passed in twenty-four hours, but without success. A legitimate inference regarding tissue metabolism cannot, therefore, be made.

An examination of the blood was made for me by Dr. A. E. Taylor, of the Pepper Clinical Laboratory and revealed the following interesting conditions. The hæmoglobin was 78 per cent. The red corpuscles numbered 4,200,000 to the cubic millimeter. In color, form, size and reaction to specific stains, these were in every way normal. The leucocytes numbered 16,000 to the cubic millimeter. A differential count of the leucocytes showed:—

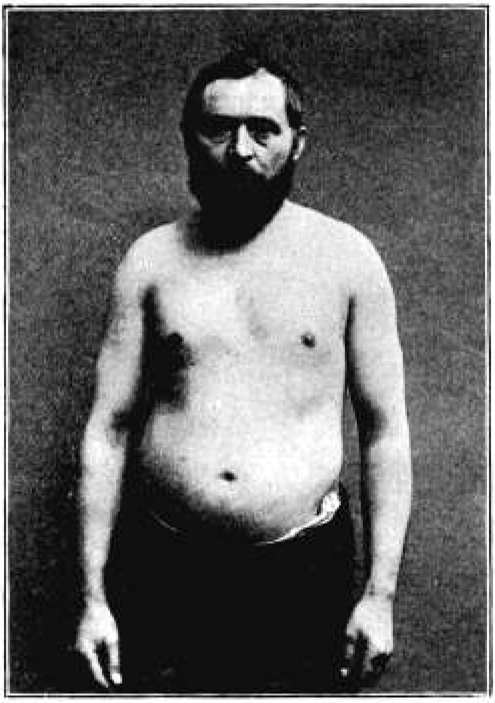

FIG. 1. C. S. Scleroderma in stage of infiltration.

Polynuclear leucocytes	56.2%
Polynuclear eosinophile leucocytes	2.0%
Simple mononuclear leucocytes	22.8%
Mononuclear eosinophile leucocytes	0 4%
Lymphocytes	18.6%

It will be observed first that the number of leucocytes taken as a whole is much in excess, and secondly that the percentage of mononuclear leucocytes is much too high. It is doubtful however whether a special significance can be attached to these findings.

The chemical examination of the blood revealed one interesting and decidedly pathological condition, namely, the presence of peptone, or rather of albumose. The fact that this substance has also been found in cases of concealed suppuration and in leukaemia makes this observation exceedingly interesting. I will not attempt to interpret its significance other than to point out how serious the modification of the normal metabolism must be to permit of its formation. As soon as the presence of peptone was discovered in the blood, I at once sought for it in the urine, but with a negative result. No peptone was found.

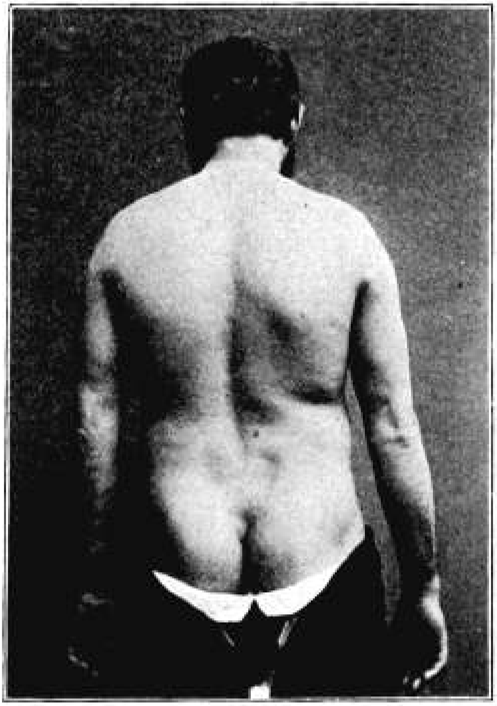

FIG. 2. As in Fig. 1. Scar over occiput is also seen.

The impression given by the photographs (see Figs. 1 and 2) is merely that of a rather stout man, though a careful examination reveals an abnormal rounding out especially of the neck and shoulders. For instance no outline of the clavicle is seen. The tissues about the eyes also appear somewhat tumid. Because of the persistent occipital headache I suggested to him that surgical interference should be undertaken which should consist of excision of the scar and possibly of exploratory trephining. This suggestion had the effect of alarming him very much and he promptly ceased his visits to the hospital and did not return until December 10, 1895.

At that time the following condition was noted:

There is present evenly diffused induration all over the neck, trunk and arms. The skin offers in these situations a firm parchment like resistance. This condition is a little less marked on the hands and face than on the trunk. As a whole the volume of the induration seems slightly less marked than at former examinations. The face seems a trifle flattened and the nose a little sharper than before. There is a diffuse reddish pigmentation all over the back. In each axilla, in each groin and in each popliteal space there is a brownish-violaceous patch of pigmentation. On the inner aspect of both arms, especially upon the left, quite a number of depressed areas are seen to which the patient himself calls attention, describing them as hollows in his arms. Upon examination it is noted that the areas of depression are more resistent than the surrounding integument and that they are also somewhat whiter. They distinctly resemble patches of morphoea save that the surrounding skin in which they are embedded is also infiltrated.

The movements of the trunk are somewhat restricted. The skin of the back grows excessively tense upon forward flexion. It is very hard and resistent. No pitting can be anywhere produced though persistent pressure with the thumb or finger appears to displace the infiltration for a short time.

Patient states that he sweats easily, that his hands and feet are frequently cold, that he is very irritable and easily excited, that he cannot do his work as well as formerly, that he gets tired earlier, and that he suffers from tinnitus.

He further states that the infiltration, especially in the hands, varies a great deal. He asserts positively that it is much more marked when he is in a warm room or when the weather is warm. He also asserts that it is more marked when he is sweating. Examination of the thyroid gland is negative. The general impression made by the examination is that the infiltration as a whole is more tense than a year ago, though the bulk of the swelling has slightly diminished. K. J. still plus. Mental depression markedly less.

The case has been examined repeatedly and of late a more or less decided recession of the enlargement and induration has been noted. On the back of the hands it has almost completely disappeared as has also the difficulty formerly experienced in extending and flexing the fingers. There is every reason to infer that the patient barely escaped the dangers of a sclerodactyle. The subsidence of the swelling appears especially about the face to be accompanied not only by a disappearance of the infiltration in the skin itself but also by a beginning loss or disappearance of the subcutaneous fat. Headache has also become markedly less.

In summing up this interesting case we are impressed first by the prominent fact presented by the etiology. It is extremely probable that the disease in this instance was to be attributed to the general nervous shock of the accident and not to the wound. The latter admitted of ready disinfection and healed rapidly and without incident. There was nothing here to suggest an infectious origin as is suggested by the well-known case reported by Touton in which a splinter was supposed to have given rise to the resulting scleroderma. In keeping with the etiological factor in this case was the persistent asthenia and mental depression. The knee-jerks which were plus presented the only noticeable physical sign of nervous disturbance and this symptom being isolated can hardly be considered as having much significance.

Evidence of neuritis, neuralgic pains and other nervous phenomena, with the exception of headache were absent in this case and the morbid process appeared to be limited to the skin and subjacent tissue.

Finally we should bear in mind the pathological condition of the blood as shown especially by the presence of peptone.

Case II, A. B., married, aged 31. One healthy child. No miscarriages. Family history is negative.

Personal History.—Beyond a history of the ordinary diseases of childhood, nothing of importance can be elicited. No history of any serious illness. Menstruated at fourteen, and was well through youth and early adult life, until the fall of 1893. At this time she suffered severely from a sudden fright, due to a slight fire in her house.

She seemed to recover from this in the course of a few days, and appeared to be entirely well until January, 1894, when it was noticed one evening that her hands seemed to be "stout and decidedly reddish." It was also remarked subsequently, by her friends, that she was looking unusually well and becoming stouter. Gradually it was noticed that the hands were more and more swollen (although the change taken as a whole was not marked). The swelling was particularly noticed in the evenings, so much so that the finger rings, which she wore, could only be removed with difficulty upon retiring. About one month after the symptoms had first been noticed the difficulty in removing the rings became so great that she had to change some of them to other fingers. These symptoms persisted until the middle of February, when it was noticed that the feet and ankles were swollen in the morning upon rising. The patient had difficulty in putting on her shoes, even old shoes that fitted loosely. At this time it was also noted that "the finger tips were spotted, and little black specks appeared upon them, giving the fingers the appearance of a person who did a large amount of hand-sewing."

About the middle of April the skin of the pulp of the finger and about the nails became very sensitive, about the nails also it became a yellowish-greenish hue. An attempt to manicure them had to be abandoned because the fingers were too sore. About this time, also, the hands were so much swollen that the patient was not able to close the hand or put on gloves.

About the end of May swelling was also noted in the legs, and there now appeared in addition a slight pain in the left wrist. Decided general weakness was also now noted, the patient not being able to walk as far as formerly. The fingers were still exceedingly sore, "being dry and scaly on the tips" Towards the middle or latter part of June she began to suffer from pain, referred especially to the elbows and wrists, and also a sense of stiffness in the neck down to the shoulders. During the summer a slight puffiness of the face and eyelids made its appearance. At the same time the skin over the bridge of the nose "became spanned or tight." The nose at this point was also extremely sensitive to the touch. During the summer her symptoms did not seem to vary much, save that the pain referred to the wrist and elbow, and the sense of tightness in the neck persisted and seemed to be more marked at times. In October there was decided increase of the pain in the wrists and elbows: it also at this time made its appearance in the knees and was very marked. The pains were now so severe, especially during the night as to interfere decidedly with sleep.

About this time the skin was swollen and stiff and somewhat reddened upon the whole of both hands and fingers, the changes being a little more marked upon the right hand than upon the left, and more pronounced upon the posterior aspects than upon the palmar surfaces. A similar condition existed in both arms gradually decreasing toward the elbows. The left side of the face presented similar symptoms, though to a slight degree. The neck seemed slightly enlarged and symmetrically so at the level of the larynx. This suggested a slight possible enlargement of the thyroid. Below the clavicle, upon the left side, there was a small area in which there was a slight stiffening of the skin. The feet and legs presented a condition resembling that seen in the hands, though to a far less degree. Motion seemed everywhere perfect except that it appeared to be interfered with by the stiffened skin, and there was no loss of sensation in any of its forms. A qualitative examination of the urine proved negative.

Her condition seemed to undergo no decided change during the following winter. At one time she had a slight swelling of the lymphatic glands under the angles of the jaws on both sides. This was accompanied by a slight febrile attack, the temperature ranging from 99 to 102.2°F. These symptoms appeared to subside in the course of a week. In December the skin in the swollen and stiffened areas seemed softer, while the joints of the limbs seemed more movable. A free perspiration was noted also at this time, the latter having, it appears, been rather scanty previously. A gradual subsidence of the swelling occurred in the early part of 1895, so that by March the face, the body, and legs seemed to have regained much of their normal appearance, though little improvement could be noted in the hands. A yellowish pigmentation appeared about this time on the finger tips. In April some pigmentation was noticed over the back and buttocks. Some improvement seemed to take place in regard to the pains in the joints and the stiffness in the neck and back.

I first saw the patient July 17, 1895. In addition to the history thus far detailed, the following fact was distinctly elicited, namely, that the changes in the surface tissues antedated the pains by six months or thereabouts. It was also learned that the pains were not limited to regions in which the skin was especially affected, nor to superficial structures, but at various times involved also the joints. The pain in the joints was described as soreness, the joints being tender to pressure, and being also at times slightly swollen. This fact was especially elicited with reference to the left elbow joint.

Her condition was as follows at the time of examination: She was considerably emaciated. Examination proved that this was largely due to an extensive loss of the superficial fat. This loss, according to the patient's statement, had followed the administration of thyroid gland. The face looked somewhat drawn, the cheeks slightly flattened, the angles of the mouth slightly drooping. When she smiled and the mouth was well opened, the libs appeared tense. The tissues of the face felt slightly more resistent to the finger than normal skin. On the right side of the face and neck a number of minute reddish areas and scales were seen, apparently the result of a dermatitis, which had suddenly supervened the previous week, and had as suddenly subsided.

A careful study of the skin of the chest showed that it was freely movable over the subcutaneous tissues. It did not seem appreciably thickened. This was also the case with the skin over the abdomen. The latter had, however, a slightly coarse feel when folded between the thumb and fingers. Its surface, also, especially over the abdomen, felt slightly uneven or irregular. On the neck, chest and abdomen it was also pigmented, especially was this the case over the abdomen, the pigment being scattered in minute patches with apparently unchanged, or slightly changed skin between. These pigmented areas suggested atrophic changes. According to the statements of the patient and of her husband, the abdomen had at one time been firm and hard, and and this had also been the case, though to a less extent, in the skin over the chest.

An examination of the back revealed the skin freely movable, not appreciably thickened, and feeling quite normal to touch. It was also pigmented or darkened, but rather in a diffuse manner, and to a much less extent than the skin of the abdomen. The skin of the buttocks was distinctly uneven to the feel, the condition being similar to that noted in the abdomen, save that it was much more pronounced. Diffuse, extensive pigmentation was also noted on the buttock, the color being of a slightly redder tint than that noted on the abdomen. In the latter region it was pale brown with perhaps a faint yellowish or greenish tinge. Over the shoulders, arms and forearms the skin was movable and not apparently thickened or changed in texture. In the lower third of the forearm, however, and especially in the neighborhood of the wrist, it felt slightly firm and resistent. Over the back of the hands, dorsal surfaces of the fingers and thumbs, it was decidedly resistent and hard. Over the smaller phalangeal joints, especially over the first joints, it was much thickened, the appearance resembling that seen in a gouty finger. In addition to being firm and resistent, the skin was smooth, shining and glossy, especially over the fingers. The hands were excessively blanched and pale, the nails were slightly ridged. The hair over the arm and forearm, however, was much coarser, darker and pronounced than normally seen in women. It was not, however, brittle. The skin, while not moist, did not feel dry. The pigmentation noted over the abdomen and other portions of the trunk, was not seen in the least degree in the hands. The arms, however, were tinged with a faint grayish-brown, resembling that seen on the trunk, but much more slightly marked.

The thighs closely resembled the upper arms. The skin was everywhere freshly movable, and was not distinctly altered to the feel save in the neighborhood of the left knee, where just above the patella a small diffuse swelling an inch and three-quarters in transverse diameter was noted. This swelling felt firm and hard, but pitted distinctly upon persistent pressure. The skin of the legs resembled that of the thighs, save that in the neighborhood of the ankles it was distinctly resistent. This was true, likewise, of the skin over the instep and behind the malleolus, and to some extent in the soles of the feet. The toes, also, appeared to be involved. The feet, like the hands, were white and glossy, though the latter feature was much less marked than the hands. The nails, as before, presented nothing unusual. The soles of the feet were slightly moist to the touch. The thighs and legs presented a discoloration similar to that seen in the arms, save that it was slightly more pronounced. As in the arms the growth of hair was more pronounced than normally. No trace of pigmentation was seen in the feet. Both hands and feet were cold to touch.

No symptoms were detected referable to the muscles, though a general wasting as from disuse or general ill health was apparent. They felt rather soft and flabby. They reacted normally to electricity. On attempting passive movements of the joints it is found that their motion was slightly restricted. This was especially true of the shoulder movements, these being restricted in upward and backward directions. This restriction could not be referred to any condition of the integument. A similar restriction was noticed on flexing the thigh upon the abdomen or on attempting abduction. Slight resistance was also met with at the elbows. At the wrist, metacarpal and phalangeal joints decided resistance was met with, both to passive and voluntary movement, the patient being both unable to extend the fingers and thumb, save to a limited degree and also unable to flex the finger and thumb more than to secure bare approximation. In the left hand even this approximation of thumb and finger was impossible. In other words a well marked sclerodactyle was present. (See Fig. 3). No tenderness over joints, muscles, nerve trunks or skin could be elicited. No loss of cutaneous sensibility in any of its forms. A small spot of redness was observed over the first joint of the little finger towards its outer aspect. The skin was glossy and hard and looked as though it were about to break down, presenting very much the appearance now and then seen in neuritis. On the helix of the right ear at its junction with the surface of the scalp was seen a small reddish surface in which some loss of tissue had evidently taken place. It suggested an ulcer which had healed.

There was also chronic conjunctivitis with loss of eye-lashes, especially those of the lower lid. She also suffered at the time from pain in the knees. This she described as dull and further stated that it was made worse by moving about. Her gait was a trifle stiff, but in no sense spastic. Movements of the trunk were much restricted, especially flexion.

The visceral examination of the patient was practically negative. She was exceedingly neurasthenic. Menstruation had not taken place for ten months; appetite and thirst were much diminished.

A few days after this examination, she was attacked by a right occipital neuralgia of great severity, the pain being with difficulty controlled. She described it as burning and tearing in character. Great local tenderness was present and soon a superficial ulcer somewhat larger than a silver dollar made its appearance low down in the right occipital region. Hair and epithelium were rapidly shed leaving a raw surface which refused to heal for a number of months. Though closely observed for a long period subsequently neuralgic attacks or attacks of neuritic pain did not occur.

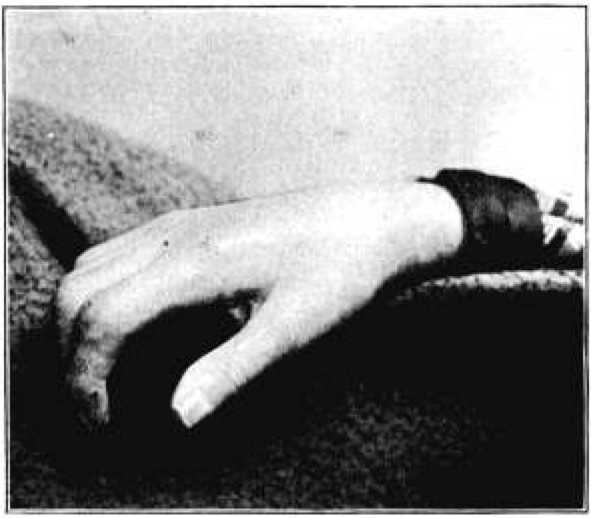

FIG. 3. Right hand of Case II.

Qualitative examination of the urine proved negative. Quantitative examination however revealed a marked diminution in the proportion of solids. The specific gravity was 1009. The total amount of urine passed in the twenty-four hours proved to be a little over forty ounces, but even assuming fifty ounces as the amount, the total solids were less than 500 grains. The diminution was especially marked as regards the urea; according to the most liberal estimate less than 275 grains were excreted daily. Uric acid was but slightly diminished if at all. This was also true of the chlorides while the phosphates were in excess.

Repeated microscopical examinations failed to reveal any evidence of renal disease. An active diuretic treatment was at once instituted, the patient taking Poland water in large quantities daily. The quantity of urine soon averaged sixty ounces daily. At first the total solids fell from 1.8 per cent. to .8 per cent. and the specific gravity to 1004. Gradually however the output of solids increased so that by December 23, 1895, it had reached 2.8 per cent. and the specific gravity 1014. This rise with slight fluctuations was maintained and at the last examination made March 25, 1896, the total solids were 3.6 per cent. while the specific gravity was 1018.

Too much importance must not be attached to these findings for the influence of food, of exercise and of other factors influencing tissue metabolism must be taken into account. We must remember that our patient at the time of the first examination was consuming but little nourishment, was taking almost no exercise and that her body weight was much diminished. It was only eighty-nine pounds. During the period that increased elimination resulted the patient was taking an increasing amount of food, was exercising to some extent in the open air and later on was being vigorously masséed. That these facts in a measure explain the findings in the urine appears extremely probable. I do not however think that they explain the latter altogether. It is certainly a very suggestive fact that along with the changes in the urine the joint pains largely subsided, that there was no recurrence of neuritis or neuritic pain and that there has been a progressive improvement in the patient's condition, not only as regards the general health but also as regards the scleroderma itself.

An examination of the blood by Dr. Taylor revealed hæmoglobin 65%; red corpuscles 4,050,000, leucocytes 10,000 per cubic millimeter; some irregularity in the red corpuscles whose color was paler than normal. A differential count of the leucocytes yielded :—

Polynuclear leucocytes	64.4%
Acidophile polynuclear leucocytes	0.8%
Mononuclear and transitional forms of leucocytes	28.0%
Lymphocytes	6.8%

As in Case 1, the porportion of mononuclear and transitional leucocytes is much too high, but qualitatively these cells are normal. A few of the leucocytes presented fragmented nuclei, signs of degeneration. The chemical examination of the blood failed to reveal albumose or peptone. I again sought for peptone in the urine but failed as in Case 1, to find it.

A number of interesting facts regarding the infiltration of the skin were noted during the progress of the case. It was observed, for instance, that the swelling above the left knee varied considerably from time to time, being sometimes much more marked than at others. At another time a streak of infiltration was noted in the left forearm several inches in length. It made its appearance in a few days, and almost as rapidly disappeared. Over the body as a whole, the pigmentation noted began gradually to lessen until at present but traces of it are left. At the same time the infiltration of the skin has grown distinctly less and less, until at present the skin has assumed an almost normal appearance over the face and extremities save in the hands and feet. Over the lower part, the forearms, wrists and backs of the hands the improvement while marked has not been as great as elsewhere. The variability noted in the swelling above the left knee was also noted in the induration of the hands. A number of times during the past three months the infiltration of the back of the hands and of the fingers was much lessened, the tissues being movable, softer and far less resistent, only to be followed by a more or less marked recurrence of the infiltration. Along with the general improvement noted in the skin, there has been an improvement in the movements of the joints, save in those of the phalanges of the hands. Here the improvement has been slight, although massage and passive movements have been faithfully practised. At my instance Roentgengraphs were made of both hands by Prof. Arthur W. Goodspeed, of the University of Pennsylvania, with the view of detecting disease of the joints or bones As is well known the cartilage of the articular surfaces is transparent to the Roentgen ray, and for this reason in the healthy hand the joints of the metacarpus and of the various phalanges reveal a clear transverse space between the ends of the bones. In these pictures, however such a space was observed only in the joints of the thumbs and in the joints formed by the metacarpal bones and the first row of phalanges. In the other phalangeal joints these clear spaces were wanting, and close examination enables us in at least one of the pictures to trace a shadow which lines the ends of the bones like a separate layer, just as though the transparent cartilage had been made opaque by the deposit of some substance not permeable to the Roentgen ray. The question arises, Have we here a mere calcareous infiltration, or an actual formation of bone to deal with? This information the rays do not give us, and practically, I fear, it makes little difference.

A steady increase in weight accompanied the improvement of the general condition and menstruation has also been re-established for three months past.

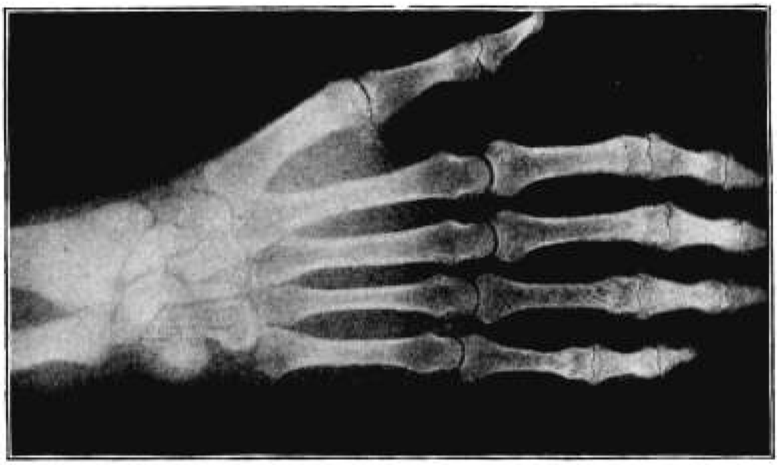

FIG. 4.

In considering Case II. a number of important points attract our attention; first, the character of the induration, which varied at times in certain places, and pitted slightly upon persistent pressure; secondly, the almost universal disappearance of the superficial fat; thirdly, the evidence in favor of the involvement of structures other than the skin and the subcutaneous tissue. Thus, the radiographs of the fingers leave no doubt of the involvement of the phalangeal joints, while regarding other joints this is a legitimate inference. For instance, about the shoulder joints the skin is soft and pliable, and has evidently completely recovered; but, notwithstanding, the movements of the joints are still much restricted. This is also the case with the movements of the trunk, flexion being more or less interfered with, although the skin of the back is soft and pliable, and can be gathered into folds between the fingers. It certainly seems as though either the ligaments of the spinal column or possibly the joints of the articular processes had been involved in the pathological process. This is true likewise of the hip joints, the restriction of movement in which can likewise not be ascribed to any condition of the skin, but depends beyond a doubt upon a change either in the joint itself or in its surrounding connective tissues, its capsular and other ligaments.

Again, the pains present were also a marked feature. They were of various kinds: first, achings in the limbs and joints independent of movement; secondly, pains referred to the joints dependent upon movement; thirdly, nerve pains, suggesting neuritis. In addition to the changes observed in the skin, various trophic changes claim our attention. They consisted of the ulceration at the base of the occiput above described, ulcerations over the phalangeal joints, the partial fixation of the fingers constituting the marked sclerodactyle, and finally the ridging of the nails.

In passing I may say that in January of this year a deep and angry looking ulcer made its appearance upon the first joint of the little finger of the right hand. At first this ulcer was simply dressed antiseptically, and kept under observation. It gradually grew deeper, while its borders widened. It was then treated by bovinine, that is the ulcer having been washed with ordinary soap and water and thoroughly rinsed, a pledget of cotton soaked with bovinine was applied and retained in position by a light bandage. This dressing was repeated twice daily. In a few days it was noted that the ulcer was becoming more shallow, and that granulations were rapidly springing up. In a short time the granulations were so profuse that the surface of the ulcer instead of being depressed was distinctly raised above the level of the surrounding skin. The local feeding with bovinine was then discontinued, and a powder consisting of boracic acid and iodol was applied in its stead, and in a short period the ulcer was well healed, without depression and covered by substantial white tissue.

Case III.[1]—[Figs. 5 and 6] M. B.: a woman; married; age 38 years; a native of Switzerland. Father died of unknown cause. Mother lived to the age of 65 years. Patient was married at the age of 22 years. Has had eight children, five of which are living. Had two miscarriages. Ten or twelve years ago suffered from malaria. Has never had a distinct attack of articular rheumatism.

Her husband being a retail ice dealer, she was in the habit of going into a small ice-house at frequent intervals during the day for a number of years.

Her present affection began about seven years ago, when she noticed that the tips of the fingers of both hands were swollen and enlarged. The tips of the fingers became clubbed, and some of them ulcerated. About three years ago the skin on the back of the hand became thick and infiltrated. This swelling did not, however, pit upon pressure. At the same time telangiectatic spots appeared upon the face. The infiltration became gradually more and more marked in the hands and fingers, and gradually interfered with their movements. Little by little the fingers became fixed in position. Over the phalangeal articulations small ulcerations occurred at various times—the result of traumatisms. Soon after the changes in the hands were noticed, swelling and infiltration appeared about the face and neck. This subsequently subsided, and left the features in the condition to be described.

Present Condition.—As the patient enters the room it is at once noted that the face is markedly altered and that there is also present a decided sclerodactyle of both hands. The skin is drawn tightly over the forehead and nose, it is shining. Numerous reddish spots are noted over the face due to enlarged capillaries. The ears are firm and unyielding. The patient cannot open her mouth as wide as formerly. The skin of the scalp is tightly adherent. The hair is somewhat thinner than formerly. Slight itching is also present. The appearance of the face is quite characteristic and the tightness of the skin and drawing of the features can readily be inferred from the photograph.

The left hand presents the following condition. The skin on the back of the hand is atrophic and tightly adherent. The index, middle and ring fingers are ankylosed at the first phalangeal articulation being fixed at almost a right angle. There is also a small ulceration on the ring-finger at this situation. The little finger is excessively atrophied being only an inch in length. This atrophy is evidently accompanied by disappearance of bone. The skin of the fingers is tightly adherent to the phalanges. The ends of the fingers are clubbed and the nails are incurvated.

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[1] For the opportunity of studying this case I am indebted to my friend, Dr. Jay F. Schamberg, of Philadelphia.

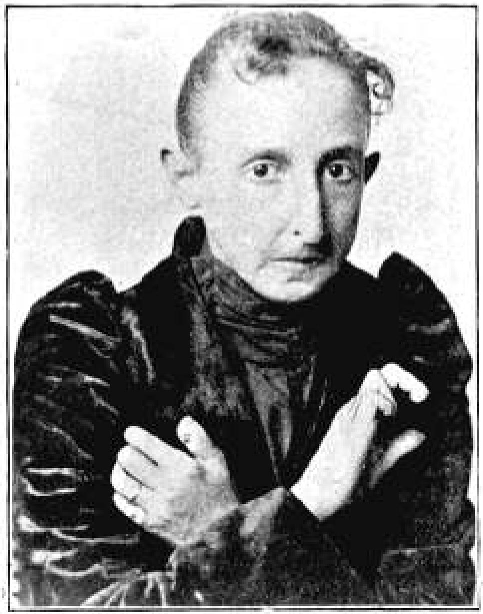


FIG. 5. Case III.

The right hand presents a condition similar to the left. All of the fingers are more or less ankylosed. In both forearms the muscles are wasted while the skin is thinned and tightly drawn. Atrophic changes are less marked in the right than in the left arm. Patient states that her maximum weight was 108, that she now weighs but 86. Superficial fat is almost entirely absent from the body. She states that she is much more sensitive to cold than formerly. She declares that the sensation of tension in skin is worse in cold weather and that the hands are at times cyanotic. The patellar reflex is minus; sensation is everywhere normal. The pupils react normally. The mucous membrane of the mouth and especially of the soft palate is unusually pale in color. Some four years ago she began losing her teeth until she was finally compelled to have them altogether replaced by a plate.

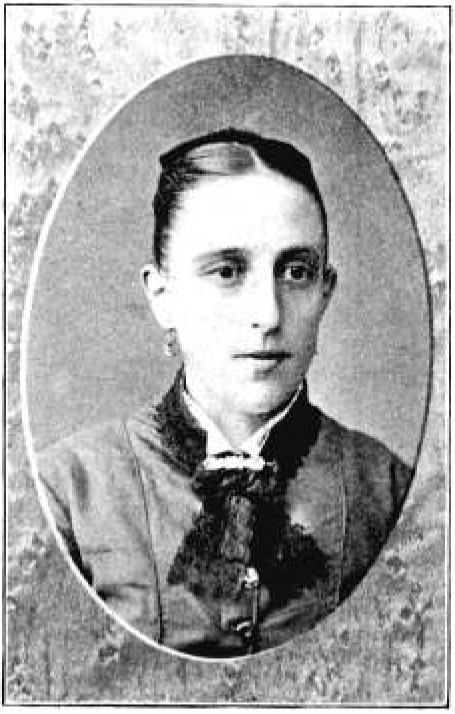


FIG. 6. Appearance of Case III. several years before onset of the disease.

In October, 1895, she was delivered of a healthy child after a rather easy labor. During the pregnancy the thyroid gland became greatly enlarged. She remained persistently weak for some time after her confinement and the telangiectatic spots over the face became much more marked than before.

The examination of the urine revealed a condition similar to that met with in Case II. The specific gravity was again only 1009. The estimation of the total output of solids was again less than 500 grains, while that of the urea was decidedly less than 200 grains. The quantity of uric acid appeared to be but slightly diminished, while the phosphates were normal. These facts probably bear here the same significance as in Case II.

In Case III the striking features are the marked atrophic changes in the skin of the face, of the forearms and hands and the very great degree of sclerodactyle. Neuralgic and other nerve pains were absent. The only nervous symptom of consequence was irregularly recurring mental depression. She has recently passed through a brief period of melancholia.

The parts of the body especially involved by the infiltration and atrophy were evidently the face, the head and upper portions of the neck, the forearms and the hands, that is just those portions which were exposed to cold in entering the refrigerator.

Before attempting to draw any inference from the cases before us and from the facts detailed, let us make a brief analysis of the symptomatology of scleroderma and determine if we can what are really the essential symptoms of the disease. When we look over the great mass of reported cases we find that the most diverse symptoms have been recorded. We find, for instance, on the side of the nervous system hypochondriasis, hysteria, depression, melancholia and mental impairment; also, insomnia, giddiness and headache and among the purely physical symptoms, chorea, tremor, ataxic movements, spasm of muscles, fibrillary twitching, local paresis, signs of intracranial lesion such as inequality of the pupils, and even symptoms suggesting Graves' Disease. Various disturbances of sensation have also been put on record; paresthesia, anæsthesia and the more common symptoms of sense of tightness and constriction. The great variety of the nervous phenomena observed, as well as the fact that they are infrequently present indicate that many of them are of an unessential character and yet the fact that nervous symptoms, though limited in number, of one kind or another are usually present indicates that the nervous system is more or less involved in this curious disease. The further fact that no one nervous symptom or group of symptoms is present in all of the cases does not militate against this inference. As showing the inconstant character of the nervous phenomena we need refer to but the reflexes as examplified in the knee-jerks. These may be normal, exaggerated or diminished.

Among the trophic changes observed are first the swelling and infiltration or induration and subsequent atrophy of the skin and subcutaneous tissues. Other and less constant trophic changes are erythematous and telangiectatic patches upon various portions of the body; pigmentation, various ulcers, small vesicles and even pustules, excessive desquamation, urticaria, and ridging, thickening and brittleness of the nails or shrinking and atrophy of these structures, Among the subdermal changes are the loss of the superficial fat and the involvement of the bones and joints, more especially of the hands. Evidently the trophic changes center primarily in the true skin and subcutaneous tissue. Changes in other tissues are as a rule not only later in onset but many of them are evidently secondary in character.

When we come to analyse the post-mortem findings, we find, first, as to distribution that the changes are by no means limited to the skin and subcutaneous tissue. As we have already seen, changes in the joints and in the tissues about them, can be inferred from many of the clinical phenomena, and is well shown in Roentgen pictures. That changes in the joints occur, particularly about the hands has been shown more especially by Legrange, who observed in sclerodactyle union of apposed joint surfaces by fibrous tissues, loss of articular cartilage and calcareous deposits in the fibrous tissues about the joints. Similar joint changes were also noted in the case studied by Verneuil and Mirault, in which short fibrous bands extended between the apposed joint surfaces, and in which there had been destruction of synovial membrane. Changes in the bones, at least of the fingers, is, of course, also at times, to be inferred from the clinical findings. Actual examination of the tissues has revealed in the hands of Wolters an interstitial inflammation of the bone, while Lagrange describes a disseminated inflammation affecting periosteum, bones and joints. Changes have also been noted in the muscles and other tissues. Foullerton. who examined the amputated leg of a case of scleroderma, infers from the widespread character of the change that scleroderma is primarily a disease of genetically related groups of tissues of mesoblastic origin; that is of the derma, the subcutaneous tissue, the muscle fibres and the fibrous capsules of the joints. Varied changes have infrequently been noted in tissues other than those here mentioned. They appear to be without special significance, and have been noted in the lungs, the heart, the liver, the peritoneum, the kidneys, the spleen, the intestine and the aorta. Cardiac changes seem to have been noted rather more frequently than others. Thus out of twenty-eight cases collected by Lewin and Heller some change, such as hypertrophy, degeneration, myocarditis or pericarditis were noted in fourteen cases.

The result of the microscopical examination of the skin in scleroderma is, of course, well known, and consists of an increase of the connective tissue elements, together with sclerotic or inflammatory changes in the blood vessels. That similar changes take place in other tissues is not only probable, but evident from the examinations which have thus far been made. It is, however, extremely doubtful to my mind whether this affection can be consideted primarily as a sclerosis. Certainly the phenomena presented by cases in which we have a recurring, or even a shifting infiltration, and which sometimes pits or yields upon persistent pressure as it did in two of the cases I have described, shows that we have every reason to believe that the changes found in the tissues microscopically are terminal changes. The clinical phenomena certainly justify the inference that primarily the connective tissue is infiltrated, swollen or enlarged. The single fact that the infiltration on the back of the hands in Case II. varied considerably from time to time, that at times the skin was soft and flexible, and at others densely infiltrated, shows that some variable or changing process not consistent with that of a primary sclerosis was at work. That a change does take place in the connective tissue other than a primary sclerosis is also held by Lewin and Heller, who regard it as very conceivable that the enlargement of the individual fibres of connective tissue depends upon their infiltration with an albuminoid substance, a saturation with some coagulable material. Unna holds that the changes are primarily in the connective tissue and especially in its intercellular or ground substance.

A view that presents itself as plausible is that the primary change in scleroderma is an increase or chemical modification of the collagen of the skin and subjacent tissues. Certainly the initial change appears to be a swelling and infiltration of the collagenic tissues. We have here a certain analogy between this disease and myxœdema. That myxœdema is accompanied by connective tissue changes, and that there is also an infiltration of the connective tissue with mucin, a substance normally present in the intercellular or ground substance, is well known, and it would seem that just as in myxœderma there is an increase of the intercellular mucin, we have in scleroderma an increase of the collagen or some, as yet, unknown chemical change in this substance, which increases the bulk of the tissues containing it. That, however, these changes differ radically from the changes in myxœdema is shown not only by a microscopic examination of the skin, by the symptomatology and by the clinical history, but also in the difference following the administration of the thyroid extract. In four cases collected by Lewin and Heller no improvement followed the administration of this remedy, so powerful in myxœdema.

Whether or not the change in the connective tissue elements is dependent upon the faulty action of some ductless gland, as is the case in myxœdema, must for the present at least, remain a matter for speculation.

It is probable that when improvement does follow the use of thyroid extract in scleroderma, that the effect is to be attributed not to action upon the swollen and infiltrated skin, but upon the subcutaneous fat, and that it materially hastens the atrophy and disappearance of this tissue in the atrophic stage of the disease.

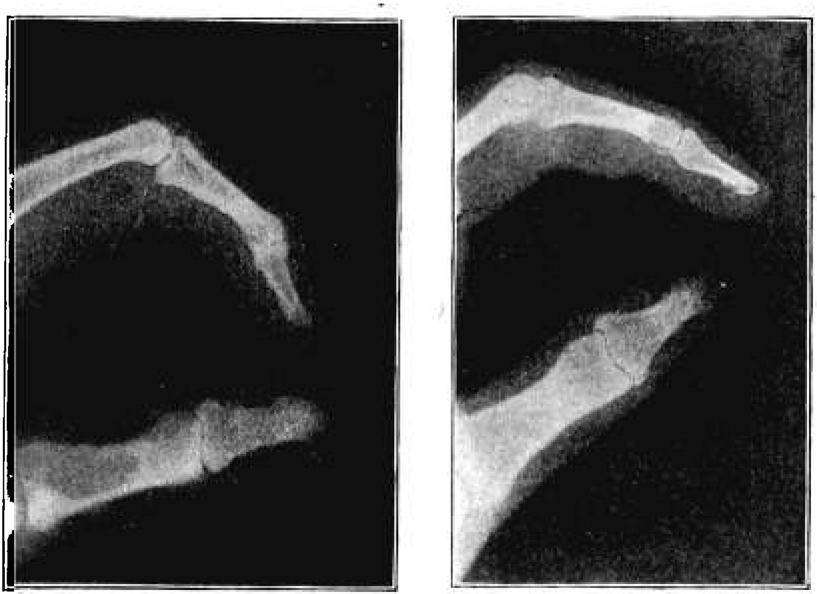
The theory that the disease is a vaso-motor neurosis, explains nothing. It merely states in a vague way that there is some relation between the changes in the skin and the condition of the central nervous system. That, however, some special, as yet unknown condition exists independent of the nervous system, is proven by the varied facts of etiology. In the first of the cases detailed here to night, for instance, a severe physical shock to the nervous system appears to have been the exciting cause. Evidently, however, such a cause could not have been the real factor at work, for in such case scleroderma instead of being a rare affection, would be an exceedingly common one. It would be as common indeed, as traumatic neurasthenia. The same is true of psychic shock, the supposed etiological factor of the second case. Certainly scleroderma must be looked upon as among the rarest of the sequelæ of psychic shock, and so it is with the cause apparently operative in the third case, repeated exposure to the cold of a refrigerator. If cold were a potential factor in the production of scleroderma, how common an affection it would be! Every winter, every climate, every avocation in which exposure to cold was attendant, would bring with it a history of scleroderma. Such a relation, it is needless to say, does not exist. Other causes which are every now and then assigned, such as muscular over-exertion, severe labor, menstrual disturbances, pre-existing infectious diseases need only to be mentioned to show how rare a sequence scleroderma is of such causes.

Whether the observations made in the cases detailed this evening in regard to the urine have any significance it will remain for future observers to determine. The number of cases is, of course, altogether too limited to permit of an inference. The facts, however, appear to be in the highest degree suggestive. The urine in Case I. (which was in the state of infiltration) suggests that an excess of waste products was being eliminated. That the urines of Cases II. and III. should have so closely resembled each other may be a mere coincidence. I doubt, however, whether this be really the case, because in Case II. distinct improvement followed, as we have seen the use of diuretic measures, and measures calculated to increase tissue metabolism. The significant fact was observed of an actual and undoubted increase of the waste products eliminated hand in hand with marked improvement. A significant fact also was the prompt subsidence of the pains in the joints.

Whether the discovery of an albumose or peptone in the blood of Case I. has a special significance is, of course, open to question. It is suggestive, however, that Case I. is in the early stage, or stage of infiltration, while Case II. is in the stage of subsidence, and it may be that this explains the difference in the findings of blood-examination.

In closing, I feel that I owe an apology for bringing to New York so scant a contribution, but my apology is first, the intense interest which trophic diseases have excited of late years, and secondly, the conviction that any facts, great or small, in so barren a field as scleroderma, must be welcome. Finally, I am not without hope that agitation and discussion of this subject may result in some practical suggestion; perhaps in a discovery which will enable us to control the disease as readily as we control that sister affection, myxœdema.

ADDENDUM.—In the discussion which followed the reading of the paper, Dr. N. E. Brill raised the point that in the semiflexed position of the fingers, such as obtains in sclerodactyle, the Röntgen rays could not pass through the phalangeal joints without encountering osseous tissue, and that therefore no clear interphalangeal spaces would be seen in the pictures. Since the reading of the paper the writer has performed another experiment with the Röntgen rays in order to determine whether this criticism holds good. It was at once learned seen that the rays, when the fingers are strongly flexed at the phalanges, do not reveal clear interspaces, even in the normal condition.



FIGS. 7 AND 8.

The writer then determined to Röntgengraph a phalangeal joint of Case II. in the *lateral* position. At the same time a Röntgengraph was made of a perfectly healthy forefinger held in the same position. Examination revealed the white line occupying the position of the cartilage already mentioned in the body of the paper, a line which seems to indicate calcareous or osseous deposit, and which is not seen in the healthy finger. (See Figs. 7 and 8.)